Chronic Wasting Disease

B. Tapscott

Factsheet

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Over the past 15 years, chronic wasting disease (CWD) has emerged as the most important disease to affect both wild and farmed cervid populations in North America. Since 2000, most jurisdictions in North America have developed surveillance programs to determine the presence or absence of CWD in both farmed and wild cervid populations. In Canada, more than 233,300 farmed and wild cervids have been tested for CWD since 1997[1]. During the same period, Ontario tested 11,074 farmed and wild cervids, and all samples tested negative. However, concerns over CWD transmission will affect the movement of live cervids and their products until better technology provides a live test with a high degree of sensitivity and specificity.

INTRODUCTION

CWD of elk and deer is an infectious disease causing animals to have increased salivation, polydypsia (increased drinking), polyuria (increased urination), excessive drooling, ataxia (incoordination) and progressive neurological signs. The minimum natural incubation period is 16–17 months, post exposure, but the maximum natural incubation period is not known. In experimentally infected cervids, incubation is known to be at least 25 months for deer and 34 months in elk[2]. CWD belongs to a group of diseases called transmissible spongiform encephalopathies (TSEs). The emergence of bovine spongiform encephalopathy (BSE) has raised the profile of CWD and other diseases in the TSE group. Hueston and Bryant have summarized the current knowledge on the TSE diseases[3].

Current research indicates that the agent associated with CWD is an abnormal infectious protein or prion. A prion is a normal cellular protein involved in synaptic (nerve junction) function at the neuron and coded by a single gene. The abnormal prion or proteinaceous infectious particle is protease resistant and abbreviated to PrP^{res}. The specific prion associated with CWD is abbreviated to Pr^{PCWD}. PrP^{res} corrupt normal cellular prions and cause them to become protease resistant. Protease-resistant prions accumulate and cause vacuolation of neurons and loss of function. A recent theory is that a slow virus is the infectious agent and the abnormal prion is a manifestation of the infection[4]. To date, there is no evidence that CWD can be transmitted naturally to other livestock or humans[5].

REGIONAL DISTRIBUTION

United States

CWD has been present in free-roaming elk, mule deer, white-tailed deer and black-tailed deer in an endemic area that includes northern Colorado, southern Wyoming and southwestern Nebraska, since the late 1970s. The prevalence rate of CWD in the endemic areas of north-central Colorado and southeastern Wyoming can be as high as 17% in local populations of mule and white-tailed deer and about 1% in elk. Wild moose from the same area have also been identified as being infected with CWD.

Many states and provinces have increased surveillance in both farmed and wild cervids. Wisconsin has tested more than 166,000 samples from wild deer[6] and in excess of 27,600 farmed cervids[7]. The common saying, "The more you test, the more you find," has unfortunately come true. Since 2001, the geographic distribution of CWD has expanded on farms and in the wild to include two Canadian provinces — Alberta and Saskatchewan — and at least 18 states — Montana, Wyoming, Utah, Colorado, New Mexico, North Dakota, South Dakota, Nebraska, Kansas, Oklahoma, Minnesota, Wisconsin, Missouri, Illinois, Michigan, New York, West Virginia and Virginia. The increased geographic distribution may be due to heightened awareness and increased testing or to a true expansion of the range of the disease.

Ministry of Agriculture, Food and Rural Affairs

Ontario

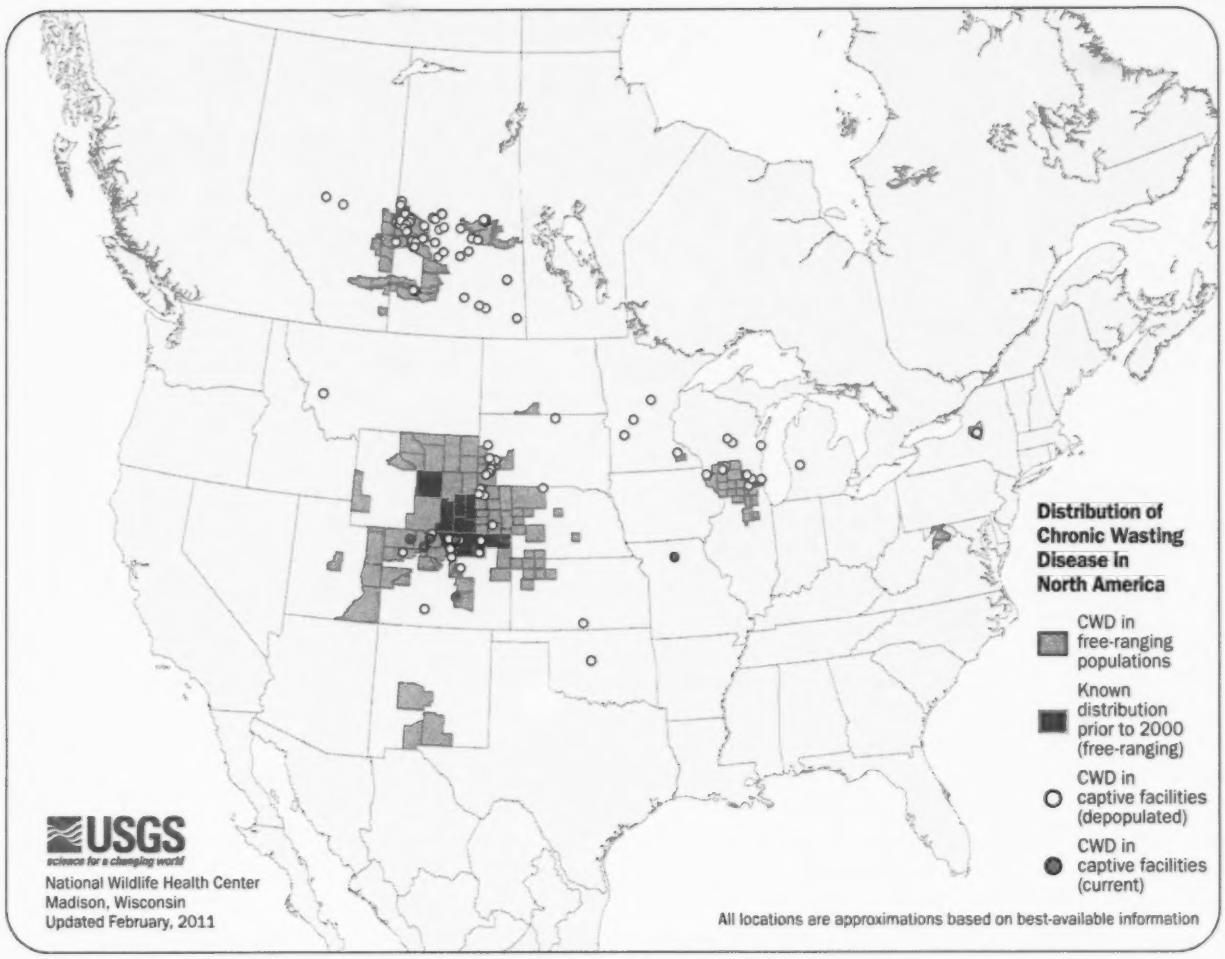
Distribution of Chronic Wasting Disease in North America
CWD in free-ranging populations
Known distribution prior to 2000 (free-ranging)
CWD in captive facilities (depopulated)
CWD in captive facilities (current)
USGS
science for a changing world
National Wildlife Health Center
Madison, Wisconsin
Updated February, 2011
All locations are approximations based on best-available information

Figure 1. Distribution of chronic wasting disease in North America, current to February 2011.

Canada

The first reported case of CWD in Canada was in a group of mule deer imported in 1974 via a shipment from the Denver Zoo to the Toronto Metropolitan Zoo. The last animal with CWD of the group died in 1981[8]. In 1996, CWD was diagnosed on 40 cervid farms in Saskatchewan. The source of this outbreak was traced to elk imported from South Dakota. This outbreak, coupled with a heightened awareness of the TSEs in the late 1990s, brought CWD to the forefront. To date, CWD has been diagnosed on 56 farms in Saskatchewan and 2 farms in Alberta.

CWD has not been diagnosed in the rest of Canada. Details of each state or province's experience with CWD are presented in the proceedings of the Second International CWD symposium[9]. The National Wildlife Health Centre summarizes CWD distribution in a map (Figure 1)[10].

TRANSMISSION

Cervid Species

Epidemiology supports CWD as primarily being an infectious disease. Maternal transmission appears to be relatively rare[5]. During the eradication of the infected farmed herds in Saskatchewan, the infection rate in herds using stock tanks with large volumes of water in the range of 270 L (60 gal) or more was greater than in herds using small-volume water bowls[11]. This would be consistent with observations that cervids exhibiting clinical signs of CWD drool while drinking and spend more time around water sources than normal. These observations support the hypothesis that saliva and feces are important in the transmission of the agent[3, 5]. The Pr^{PCWD} accumulates in gut-associated lymphoid tissue (e.g., tonsils, mesenteric lymph nodes). Saliva and feces are important in the shedding of the agent.

TSE infectivity can persist, when buried, for at least 3 years[12]. Practices that increase the concentration of cervids and, therefore, the environmental contamination (e.g., holding cervids in captivity or through artificial baiting of wild cervids) may be important in increasing the transmission of the agent. Casual fence-line contact is less likely to increase the possibility of transmission. Both direct (e.g., prolonged contact across the fence) and indirect transmission (e.g., from contaminated pastures) are possible. Shedding of the virus probably precedes clinical signs in both elk and deer[5].

In the Saskatchewan eradication of farmed elk, genetic analysis showed that the majority of CWD test-positive elk were homozygous for 132M. CWD was not detected in elk that were homozygous for 132L[13].

Wild Cervids

In free-roaming mule deer, adult males may play an important role in the spread of CWD. The prevalence of CWD peaks in 5–6-year-old males and shows no age-related pattern in females. Gender-related behaviour, such as the roaming of sexually mature males during breeding season, with increased opportunities to interact with sources of infection and associate with numerous females, might explain the differences in infection rate (prevalence) between genders[14].

Domestic Livestock

Cattle, sheep and goats seem to be relatively resistant to infection. A long-term study intensively exposed cattle to CWD-infected deer and elk via oral inoculation or confinement with infected captive mule deer and elk. The cattle remained healthy for over 5 years[5].

Humans

On occasion, media reports have indicated that hunters have died of Creutzfeldt-Jakob Disease (CJD). Investigations into these reports have not supported their claims[15]. Transgenic mice expressing the elk prion protein — "cervidized mice" — were intracerebrally inoculated with elk CWD prion. Two genetic lines of "humanized" transgenic mice that were susceptible to human prions (PrP^{res}) were also intracerebrally inoculated with elk CWD prion. The humanized mice failed to develop CWD after more than 657 and more than 756 days, respectively. In contrast, the "cervidized" transgenic mice became infected after 118–142 days. This data may indicate that there is a substantial species barrier for transmission of elk CWD to humans[16].

SURVEILLANCE

Wild Cervids

Many jurisdictions have developed surveillance programs for wild cervids using sampling frameworks based on geographic randomized sampling, using low prevalence rates (less than 1%) and probabilities of detecting the first case at or above the 95% confidence level. Various jurisdictions target their surveillance to areas based on perceived relative risk. Factors, such as proximity to provinces or states with CWD or areas where cervid farms are concentrated, are used to prioritize surveillance. Hunter-killed cervids are a common source of surveillance samples. In some areas, especially if there is no hunting season, road-kill submissions are used for CWD surveillance. The Ontario Ministry of Natural Resources has tested over 8,533 wild cervids, with no positive cases to date (Table 1).

Farmed Cervids

Alberta, Saskatchewan and Manitoba currently have mandatory surveillance programs. All deaths and slaughter animals greater than 12 months of age must be tested for CWD. Quebec and Ontario have voluntary surveillance programs. The remaining provinces have very few or no farmed cervids. CWD is a reportable disease in Canada, where all suspect cases must be reported to the Canadian Food Inspection Agency (CFIA) under the *Health of Animals Act.*

Ontario has tested 2,541 farmed cervids since 1998, and all samples have tested negative to date (Table 1).

In the U.S., a federal CWD herd certification program is in the development stages. Many states have placed a moratorium on the import of cervids, both farmed and wild, into their state. CWD surveillance programs vary from state to state; some programs are voluntary, and some are mandatory. The CFIA and U.S. Department of Agriculture are discussing harmonization of Canadian and U.S. CWD herd certification programs.

Table 1. Provincial CWD sample submissions for Ontario, 1998–2010

Year	Farmed/Captive							Wild	Total Farmed + Wild
	Elk	White-Tailed Deer	Red Deer	Fallow Deer	Reindeer	Unspecified	Total Farmed/Captive		
2010	102	49	129	19	1	0	300	1,401	1,701
2009	156	98	24	0	0	0	278	771	1,049
2008	149	94	97	5	2	0	347	1,549	1,896
2007	200	91	63	20	1	0	375	882	1,257
2006	194	76	21	0	0	25	316	1,438	1,754
2005	41	43	16	1		9	110	1,411	1,521
2004	115	52	23	1		43	234	427	661
2003	167	47	16	1		10	241	471	712
2002						121	121	183	304
2001						71	71		71
2000						50	50		50
1999	16	9	15	3		4	47		47
1998	15	10	18	6		2	51		51
Total	**1,155**	**569**	**422**	**56**	**4**	**335**	**2,541**	**8,533**	**11,074**

SAMPLE COLLECTION AND TESTING

From the late 1990s until the fall of 2005, diagnosis of CWD was made by the presence or absence of the abnormal protein PrP^{res} on immunohistochemistry (IHC). IHC is referred to as the gold standard for CWD diagnosis. The obex section of the brain stem (*medulla oblongata*) is the preferred site (Figure 2). It is collected and placed in 10% buffered formalin. Pathologists evaluate the staining in the region of the parasympathetic vagal nuclei.

More recently, "rapid tests" have been developed and approved for use. Most of these rapid tests are ELISA based, while one is a commercial Western blot assay. Fresh or frozen samples are submitted to the laboratory, and results are often available within 24–36 hours. The Bio-Rad ELISA test is the most widely used for CWD testing in Canada. Since 2004, the retropharyngeal lymph nodes (RPLN) have been the preferred tissue for testing in white-tailed and mule deer, while the obex remains the preferred tissue in elk and red deer[13]. Biopsy and testing of tonsillar material has been tried[17]. Canadian researchers are currently evaluating the use of rectal mucosa as a live cervid test[18]. Researchers at the University of Guelph have been experimenting with an acoustic sensor for prion detection in blood and urine[19].

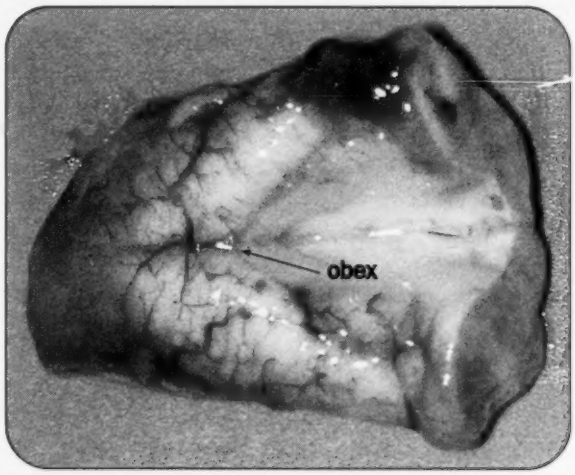

Figure 2. Cut the brain stem 1 cm (0.4 in.) above and below the obex (the "V" on the dorsal surface of the brain stem).

For the Bio-Rad test, place the obex and retropharyngeal lymph nodes (RPLN) in separate whirl-packs and refrigerate or freeze the tissues prior to submitting to the laboratory.

For the immunohistochemistry (IHC) test, place the obex/RPLN in 10% buffered formalin. Some jurisdictions may require samples to be split, with half placed in buffered formalin and half fresh or frozen.

The Animal Health Laboratory (AHL), at the University of Guelph, is now using Bio-Rad to test for CWD and is no longer using IHC. All samples from Ontario to be tested using Bio-Rad must be shipped fresh or fresh/frozen. Ontario samples can be shipped to the AHL by Purolator at no cost, by marking "Purolator — University of Guelph incoming account #096691" on the waybill. Samples shipped in formalin can no longer be tested by AHL.

SAMPLING AT ABATTOIRS

When developing surveillance programs of slaughter animals, it is necessary to hold carcasses until the samples have been declared free of CWD. The turn-around time between tissue collection, shipment, testing and release of carcasses can be a problem when cooler capacity in an abattoir is limited. This is less of a problem with the new rapid tests that have turnaround times of 24–36 hours, but still requires the coordination of meat inspection and laboratory services.

SUMMARY

With the detection of CWD in 18 states and 2 provinces, its long incubation period, and the public's heightened awareness and fear of the TSE diseases, it is critical that cervid farmers and governments continue surveillance to halt the spread of CWD.

This Factsheet was updated by Brian Tapscott, Alternative Livestock Specialist, OMAFRA, Elora. The original Factsheet was authored by Brian Tapscott, and Dr. Bob Wright, Veterinarian — Disease Prevention, Equine and Alternate Species, OMAFRA, retired.

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FOR YOUR NOTES

Agricultural Information Contact Centre:
1-877-424-1300
E-mail: ag.info.omafra@ontario.ca
Northern Ontario Regional Office:
1-800-461-6132

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